

## EDITORIAL COMMENT

# Surgery for Bicuspid Aortopathy

## Precision Imaging to Time Decision-Making\*



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*One accurate measurement is worth  
a thousand expert opinions*

—Grace Hopper,  
U.S. Navy Rear Admiral

**B**icuspid aortic valve (BAV) is a common congenital cardiac abnormality, affecting approximately 1% to 2% of adults,<sup>1</sup> with a high prevalence (20%-80%) of concomitant aortopathy.<sup>2-10</sup> The primary concern associated with aortopathy remains progressive aortic dilation with risk of dissection or rupture. Despite improvements in diagnostic and surgical techniques, the mortality of acute type A aortic dissection (ATAD) remains high, ranging from 14% to 30%.<sup>11-13</sup> Hence, preemptive aortic surgery is recommended in BAV patients whose aortic dimensions reach a certain threshold. Indeed, in patients with appropriately timed aortic surgery, the incidence of aortic rupture and/or dissection is very low, with a survival similar to that of an age- and gender-matched normal population.<sup>10</sup>

Prior observations have demonstrated that ~40% of patients have aortic diameters <5.0 cm at the time of ATAD.<sup>14</sup> Similarly, a recent paper has suggested that in unoperated thoracic aortic aneurysm (TAA) patients (~9% with BAV), an aortic size of 5 cm rather than 5.5 cm was a more appropriate intervention criterion for prophylactic surgery.<sup>15</sup> Another study has suggested that the aortic size cutoff of 5 cm should be

similarly applied to BAV and trileaflet aortic valve patients.<sup>16</sup> Indeed, the most recent guidelines recommend prophylactic TAA surgery in BAV patients with an aortic size  $\geq 5$  cm, especially with risk factors and if performed in an experienced high-volume center.<sup>17</sup>

In this issue of *JACC: Advances*, in the study from 7 centers (1 U.S., 5 European, and 1 Argentinian), 875 BAV patients with ascending aorta 5 to 5.4 cm under surveillance, Ye<sup>18</sup> reports that the incidence of ATAD is low and the overall rates of ATAD and surgical mortality are similar, suggesting clinical equivalence between surgical and surveillance strategies. There are obvious limitations in the study, rightfully acknowledged by the authors. Most patients (84%) came from a single tertiary U.S. system where majority of the patients get referred from relatively long distances. Also, mostly echocardiographic measurements were used, which were site-reported rather than core laboratory-measured.

We must evolve in terms of precision imaging. Application of an unindexed unidimensional measure on echocardiography (eg, aortic diameter) fails to recognize 2 important factors: 1) body size is a significant determinant of normal aortic dimension; and 2) a 2-dimensional measure of a 3-dimensional structure like the aorta may not fully account for eccentricity. Two-dimensional echocardiograms may result in oblique cross-sections through the aorta, leading to under- or over-estimation of aortic size. As a result, we cannot and should not be making prognostic predictions using imprecise data obtained on echocardiography. To accurately determine the largest diameter, double-oblique cross-sections through the aorta are recommended to generate aortic cross-sectional area. It is important to perform electrocardiographic-triggered gated computed tomography or magnetic resonance angiography when measuring aortic dimensions, and measurements should not be used interchangeably across

\*Editorials published in *JACC: Advances* reflect the views of the authors and do not necessarily represent the views of *JACC: Advances* or the American College of Cardiology.

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modalities. Also, body size should be accounted for when making aortic measurements. We believe that aortic cross-sectional area-to-height (CSAH) ratio is the optimal technique for quantifying risk of future aortic complications, especially in the setting of eccentrically dilated roots. Indeed, a root CSAH of  $>10 \text{ cm}^2/\text{m}$  has been shown to correlate with higher rates of death, providing improved risk stratification, including in patients with BAV or trileaflet aortic valves and concomitant aortopathy.<sup>19,20</sup> In the most recent guidelines, prophylactic thoracic aortic surgery is recommended at CSAH ratio  $\geq 10 \text{ cm}^2/\text{m}$ .<sup>17</sup> It is also important to note that most patients with aortic sizes between 5.0 and 5.5 cm will transition to surgery at some time if they do not die before surgery. Thus, it is incumbent on physicians to follow these patients closely for subtle changes using precise imaging if they are not referred for surgery.

Certainly, lowering current surgical thresholds should be balanced against the downstream consequences, which would potentially expose many people to a major operation that has widely varied outcomes and complication rates. Decisions to intervene earlier should consider precise aortic measurements, local surgical outcomes, ability to offer comprehensive aortic valve/aortic care, and patient preferences. For instance, prophylactic valve-sparing root replacement with aortic valve repair at a center of excellence can be achieved with 0.16% mortality, better than 95% 10-year freedom from reoperation, and downstream descending aortic dissection of 1.6%.<sup>21</sup> These numbers are significantly better than what has been described in the current paper.<sup>18</sup>

Finally, as a society, we need to significantly improve care of patients with ATAD. Unfortunately, even ATAD patients treated at many high-volume academic centers have an average mortality ranging from 14% to 17%,<sup>22,23</sup> while quaternary aortic centers can have mortality  $<8\%$ .<sup>24</sup> However, access to such quaternary centers is often limited and must be balanced with expeditiousness of care delivery.

In conclusion, given the vastly improved outcomes of TAA repair, accurate timing of prophylactic intervention is crucial. Current guidelines are evolving toward an earlier TAA operation given recent data showing a higher rate of complications in patients with aortic diameters below recommended thresholds. However, it is crucial to focus on precision imaging to guide precise timing of surgery. Simultaneously, our bigger calling is to create high-volume centers where such patients can be treated expeditiously and safely with the best outcomes using the widest spectrum of modern procedures.

#### FUNDING SUPPORT AND AUTHOR DISCLOSURES

Dr Desai is the Haslam Family Endowed Chair in Cardiovascular Medicine. Dr Svensson is the Delos M. Cosgrove, MD, Chair for Heart Disease Research. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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**KEY WORDS** aortic dissection, aortic size, outcomes